Right Coronary Artery Fistula to Left Ventricle Complicated with Huge Coronary Artery Aneurysm

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Abstract

Congenital coronary artery fistula (CAF) with huge coronary artery aneurysm is a very rare condition. In this paper, we describe a 26-year-old asymptomatic male patient with right coronary artery (RCA) to the left ventricle fistula with a huge coronary artery aneurysm which was diagnosed by multidetector computed tomography and coronary angiography. The patient received surgical treatment for coronary artery after diagnosis. Both RCA and a giant aneurysm were excised; surgical closure of CAF and coronary artery bypass grafting were performed on this patient. Two months after surgery, the enlarged left ventricle returned to normal as evaluated by echocardiography.

Key words: coronary artery fistula, coronary artery aneurysm, multidetector computed tomography, surgical treatment


Introduction

Coronary artery fistula (CAF), which is defined as the abnormal direct communication of a coronary artery vessel with a cardiac chamber, a great vessel or another vascular structure bypassing the capillary bed, is a relatively rare condition, occurring in 0.1% to 0.2% of coronary angiographic studies (1). Low pressure structures are the most common sites of CAF drainage and only in very rare instances is termination of the coronary artery fistula in a left heart chamber (2, 3). The association of CAF with a huge coronary artery aneurysm is even more rare (4). In this paper, to provide a better understanding of this disease, we describe a case of right coronary artery fistula to the left ventricle with a huge coronary artery aneurysm.

Case Report

A 26-year-old man was admitted for heart murmurs and cardiomegaly detected by X-ray film during a routine physical examination. He did not present any chief complaint. No previous similar medical history was found in his family. On physical examination, he had normal development and respiration. Blood pressure was 130/50 mmHg. Chest examination revealed moderate to severe whiffing diastolic murmurs on apex without any conduction. The heart shadow was enlarged towards the left side. Traube’s sign was auscultated on the bilateral femoral artery and Quincke’s sign could also be detected. Echocardiography revealed enlargement of the right coronary artery, with a 23 mm inner diameter, which drained into the posterior wall of the left ventricle (Fig. 1). The left ventricle was obviously dilated (71 mm) with a normal ejection fraction (EF) of the left ventricle (65%). Electrocardiography showed normal sinus rhythm with 67 beats per minute. The coronary angiography showed enlargement of the right coronary artery (RCA) with a distal aneurysm which protruded into the left ventricle (Fig. 2); the left coronary artery and branches appeared normal. To obtain precise information on the anatomy and termination of the fistula, a 64-slice multidetector computed tomography (MDCT) angiogram was...
performed. The results showed the fistula arising from the right coronary artery and emptying into the dilated left ventricle (Fig. 3a and 3b) and a giant aneurysm in the right coronary artery was formed at the entrance of the fistula drainage site with a maximum diameter of 58 mm (Fig. 3c and 3d).

The patient received surgical treatment for coronary artery after diagnosis. The great saphenous vein graft was then anastomosed from the aortic root to the posterior descending artery and the acute marginal branch of the RCA. Both the RCA and giant aneurysm were excised. A 2.0 cm² Dacron Patch was used to close the orifice of RCA. MDCT after surgery showed absence of RCA and the aneurysm and in its place, the saphenous vein graft (Fig. 3e-3h). The surgically excised segment of the RCA showed a large aneurysm.

The pathologic examination after surgery validated the diagnosis of coronary artery aneurysm. The wall of the RCA was characterized with marked thinning of the media and abnormal arrangement of muscle fiber (Fig. 4a); red blood cells and plaques could be easily identified in the arteriole (Fig. 4b).

The patient presented no symptoms after surgery. Traube’s sign, Corrigan’s pulse and Quincke’s sign were all absent after surgery. The dilated left ventricle inner diameter retracted to within 50 mm after 60 days by echocardiography with a normal EF. He is currently still being followed-up with no reported complaints.

**Discussion**

While congenital coronary anomalies occur in 1% to 2% of the population, coronary artery fistulae (CAF) account for only 0.2 to 0.4% of all patients in previous studies of congenital heart disease (CHD) (5, 6), producing an estimate of 0.002% for the general population (5-8). The etiology of CAFs, as in the present case, is most frequently congenital. Around 55%-65% of congenital coronary artery fistulae arise from the right coronary artery and usually drain into a right chamber (9); right coronary artery into a left chamber is less frequent. To our best knowledge, only a few reports in the English language literature have been associated with RCA fistula drainage into the left ventricle (10-12). The as-
association of CAF with coronary artery aneurysm is even more rare (4). This aneurysm is regarded as one of the complications of CAF, where the RCA communicates with the left ventricle, as published in a few English language reports (13, 14). Most of the patients with CAF older than 20 years, although they may remain asymptomatic as in this case, develop symptoms, like angina pectoris (14, 15) and infective endocarditis (16), with fistula-related complications with increasing age. Mortality and morbidity is hence increased when surgery is performed in later life (17). Although patients can be asymptomatic, they can have notable abnormalities on radiography or echocardiography as reported in the present case, and heart failure would have been a likely consequence for this patient if the anatomic abnormality had not been appropriately corrected.

Until recently, conventional coronary angiography was still the diagnostic method of choice for detecting coronary anomalies. While the coronary angiography may show the

Figure 3. Results of multidetector computed tomography angiography. Three-dimensional volume rendered images of MDCT showed preoperative and postoperative reconstructive image of vessel anatomy and heart. From a to d, the surface anatomy of RCA (right coronary artery) and coronary artery aneurysm were clearly visualized. The RCA was dilated and tortuous with direct protrusion into the left ventricle (a-b). In the distal portion of the RCA, a giant coronary artery aneurysm (maximum diameter: 58mm) was formed without showing distal branching at the entrance site of left ventricle (c-d). After surgery, both RCA and giant aneurysm were excised instead of the great saphenous vein graft (red arrows) which was anastomosed from the aortic root to the posterior descending artery and acute marginal branch of RCA (e-g). The isolated vascular structures clearly showed the anatomy of the vessels (h). (AO: aorta, AS: aortic sinus, RV: right ventricle, RCA: right coronary artery, LV: left ventricle, GA: giant aneurysm)

Figure 4. Histological images of the RCA wall. The wall of the RCA showed a marked thinning of the media and abnormal arrangement of muscle fibers (a, ×400); extensive elastic tissue deposition can be identified within the medial wall and collagen tissue. Higher magnification showed red blood cells and plaques (arrows) that could be found in the arteriole (b, ×1000).
origin of fistula, the course and drainage site may be not so clearly visualized (18). Further developments have enabled coronary arteries to be evaluated using MDCT (19). Although most fistulae are diagnosed with transthoracic echocardiography and coronary angiography, the present case demonstrates the utility of multi-slice CTA with coronary anomalies which provided the exact drainage pathway. Moreover, it is not only an easy and reliable non-invasive imaging technique to confirm the diagnosis and surgical plan for treatment, but it provides far better spatial resolution than any other noninvasive imaging modality and allows volumetric (multidimension) reconstruction as well (12). Although the radiation exposure associated with MDCT can be high, the use of dose-reducing strategies can make this method acceptable, such as presented here (20).

Closure of the congenital coronary artery fistula is widely recommended in symptomatic patients (21), but is still controversial in patients without symptoms (22). Some research recommends closure of congenital coronary artery fistulae, even in asymptomatic patients in order to prevent fistula-related complications with increasing age, especially because of the risk of endocarditis (21). Although the present patient was still asymptomatic, his left ventricle was so large that any further enlargement might have led to cardiac output insufficiency. In addition, the risk of a giant aneurysm rupture and endocarditis can also not be neglected. Based on the above careful considerations, we decided to undertake surgery. Subsequently, the patient’s Traube’s sign, Corrigan’s pulse and Quincke’s sign were all absent after surgery, likely due to the hemodynamic changes in the coronary artery. On the other hand, it has also been reported that some cases may not require immediate treatment and these patients should be carefully monitored for any changes (23).

In conclusion, RCA with left ventricle drainage and giant coronary artery aneurysm are rarely seen in congenital CAF. MDCT is a very useful tool in the diagnosis of CAFs. The optimal treatment for coronary artery fistula with giant coronary aneurysm should be coronary surgery, even in the asymptomatic patient.

The authors state that they have no Conflict of Interest (COI).

References